308 Change in prevalence of liver parenchyma ultrasound abnormalities in children with cystic fibrosis

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Background: The prevalence of liver steatosis in patients with CF is reported to be 23–67%. These estimates are from histopathological studies performed following clinical or biochemical indication of hepatic abnormality. The ability of ultrasound scan (USS) of liver parenchyma to differentiate features consistent with fatty infiltration (increased reflectivity) from fibrosis (heterogeneity) has long been established.
In 1996 we reported a cross sectional survey of liver USS in 85 patients, age mean(±SD) = 11.14(±4.1) years. Fatty liver was seen in 3 patients and heterogeneous parenchyma in 28 (33%).
Aim: To review the liver USS findings in our current paediatric CF clinic.
Methods: Every child over 5 years old has liver USS performed as part of annual assessment by one of a limited number of experienced sonographers. Fatty liver was defined as increased reflectivity of the liver parenchyma. All assessments performed in 2009 were reviewed.
Results: Liver USS was performed on 90 patients mean age(±SD) = 11.84(±3.44) years. The results are presented in the table.

	Liver USS parenchyma		
	Normal (N = 62, 68.9%)	Fatty (N = 25, 27.8%)	Heterogeneous (N = 3, 3.3%)
Male	33 (53.2%)	7 (28%)	3 (100%)
Age (y), mean±SD	11.43±3.55	12.63±2.75	13.81±5.12
BMI centile, median (range)	51.2 (0.3–94.7)	47 (15.9–89.6)	21.2 (8–75)
Homozygous DF508	42 (67.7%)	14 (56%)	1 (33.3%)
CFRD	2 (3.2%)	4 (16%)	0
Pancreatic insufficiency	61 (98.4%)	24 (96%)	3 (100%)

Conclusion: Compared to 1996 USS diagnosed fatty liver has become more common with a decrease in fibrosis. CFRD and female sex were more prevalent in the patients with fatty liver on USS. As the survival of people with CF continues to increase it is essential to determine the significance of this non-alcoholic fatty liver disease

309 Evaluation of a new ultrasound scoring system for CF liver disease

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Background: CF liver disease (CFLD) is common among patients with CF with an incidence of 10%. It is a major cause of morbidity and mortality. Early diagnosis of CFLD is difficult and often missed until advanced liver disease and portal hypertension are present. The aim of this study is to implement the Stuart Ultrasound Scoring System (SUSS), a new Ultrasound scoring system, in children with CF, to assess its ability to diagnose CFLD and consider early intervention.
Methods: The SUSS protocol includes evaluation of: liver edge and texture, size of spleen, portal tracts and hepatic vein waveform. These parameters were scored either as normal (1) or abnormal (2). In addition, the liver was categorized according to the general appearance as normal (A), bright or echogenic (B) and granular (C). A normal score is A5, and any other score is abnormal. Furthermore, portal vein and hepatic artery and vein diameter were measured. Investigations included AST, ALT, GGT, albumin and INR.
Results: Ultrasound was performed on 22 patients (aged 2 to 34 years old) with CF. Two of them have known CFLD. 15 had normal score (A5) and 7 patients (33%) had abnormal scores ranging from A8 to C10; including the patients with CFLD but the rest were not thought to have liver disease previously. There was no correlation between the SUSS score and biochemical liver function.
Summary and Conclusions: This preliminary study assessed and verified the use of a novel US scoring system for CFLD. It detected previously undiagnosed ultrasonographic liver abnormalities. These results support the need for further evaluation of this score with liver biopsy confirmation and long term follow-up.

310* Liver elastography to detect early stage cystic fibrosis related liver disease in children

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Background: Cystic fibrosis related liver disease (CFLD) is an important cause of mortality and morbidity in CF patients. Unfortunately sensitive diagnostic tools to detect the early stages of CFLD are lacking, thus the diagnosis is usually only made when advanced disease is present.
Aim: Elastography of the liver has proven to be a valuable tool to detect the early stages of permanent liver damage in other liver diseases. This study therefore aimed to evaluate the value of elastography in predicting early stages of CFLD.
Methods: Between January 2009 and January 2010, 100 children with CF were prospectively included (age 4 to 18 yrs). Liver stiffness was measured by elastography (Fibroscan, Echosens, Paris) with the pediatric probe, and compared to abdominal ultrasound (US) and serum ASAT, ALAT and GGT data. Results were compared between patients with (1) normal liver tests and US, (2) early stage liver disease (abnormal ASAT, ALAT, GGT or mild US abnormalities, i.e. liver irregularity) and (3) severe CFLD (incl. nodular liver edge and/or splenomegaly).
Results: Valid measurements (IQR < 30%, success rate >70%) were obtained for 86%. In comparison to CF patients without any liver pathology, who had a mean liver stiffness of 4.2±1.0 kPa, both patients with early stage liver disease and patients with CFLD had a significantly higher mean liver stiffness (6.2±4.5; $p = 0.015$ and 9.2±5.3 kPa; $p < 0.001$, respectively).
Conclusions: Liver elastography is a useful tool for the detection of early stage liver disease in CF patients. Elastography might also be valuable to evaluate the effectiveness of ursodeoxycholic acid in the treatment of CFLD, which is still uncertain.

311 Plasma advanced oxidation protein products in cystic fibrosis patients with liver disease

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Background: Although the pathogenic mechanism underlying liver cirrhosis in cystic fibrosis is still unclear, a role of oxidative stress in this disease has been suggested. Advanced oxidation protein products (AOPP) are markers of oxidant-mediated protein damage. It has been established that the liver and the spleen play important roles in the elimination of AOPP. Liver cirrhosis can decrease AOPP plasma clearance that can lead to increase its plasmatic level.
Aim: To assess whether serum advanced oxidation protein products (AOPP) concentrations are abnormal in CF patients with liver cirrhosis.
Methods: A total of 30 children with CF were included in the study. We analyzed AOPP in the serum, comparing CF patients with liver cirrhosis (n = 12, mean age: 10.5 years, range 3.5–15) and without it (n = 18, mean age: 10.8 years, range 6–14).
Results: The plasma content of AOPP found in CF patients with liver cirrhosis was significantly higher compared to patients without it (56.46±7.93 and 38.3±2.55 μmol/L, mean±SEM, $p = 0.022$), respectively.
Conclusions: Plasma concentration of AOPP were significantly increased in CF children with clinical, biochemical and ultrasound evidence of liver cirrhosis. Measurement of AOPP concentration may be useful to reveal severe liver disease in CF patients.